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Two center study to assess the functional relevance of myocardial fibrosis in muscular dystrophy patients with and without left ventricular systolic dysfunction

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Background

Abnormalities of dystrophin expression cause Duchenne Muscular Dystrophy (D) or Becker Muscular Dystrophy (B). Since widespread use of non-invasive respiratory ventilation, many D patients who previously died in their early 20 s from respiratory failure are now surviving longer and developing cardiomyopathy. In B patients, disease progression is slower, however, development of progressive cardiomyopathy is highly frequent. One previously identified predictor of mortality in cardiomyopathy is myocardial fibrosis as detected by late gadolinium enhacement (LGE) cardiovascular magnetic resonance (CMR) imaging. Furthermore, a high percentage and typical inferolateral pattern of LGE has been previously reported in small numbers of patients with B/D, however, the relationship with left ventricular systolic dysfunction needs to be further evaluated.

Methods and Results

In this two center study, 25 patients (median age 40 yrs, range 11 to 60) with skeletal muscle biopsy and/or genetically detected dystrophin abnormalities were prospectively recruited from the Division of Cardiology of the Robert-Bosch-Krankenhaus, Stuttgart, Germany and the John Radcliffe Hospital, Oxford, UK. CMR was performed using either a Siemens Magnetom Sonata 1.5-T (Erlangen,

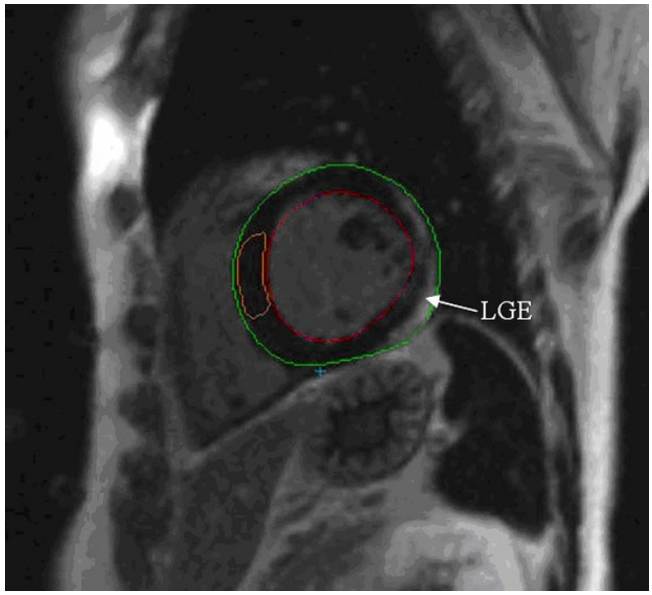

Figure 1
Short axis slice showing LGE in the lateral and inferolateral walls. The unaffected reion in the septum provided the reference pont for LGE quantitative analysis.

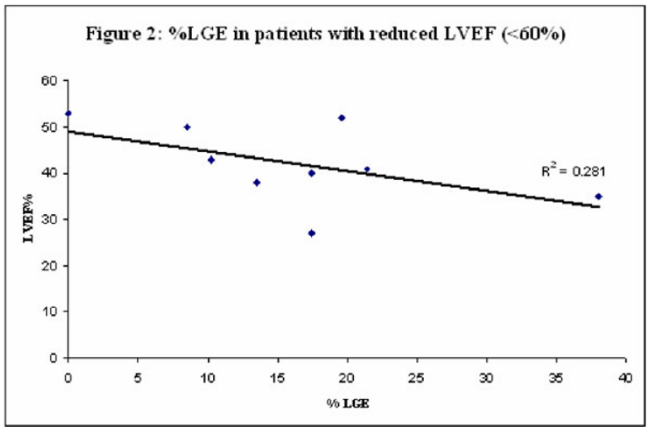

Figure 2
%LGE in patients with reduced LVEF (< 60%).

Germany) or 3-T Siemens Tim Trio (Erlangen, Germany). Anatomical, functional and LGE imaging with quantification of LGE extent (Figure 1) were performed.

Median left ventricular ejection fraction (LVEF) was 62% (range 27 to 71%). LVEF was normal (? 60%) in 14/25 patients (median LVEF 66%, range 61 to 71%) and reduced in 11/25 patients (median LVEF 43%, range 27 to 57%). LGE was present in 19/25 patients (median LGE/total mass 17%, range 0 to 38%). Presence of LGE was observed in 10/11 patients with reduced but also in 9/14 patients with normal LVEF (p = 0.60). The median size of total LGE was higher for patients with reduced LVEF (median LGE 17.4%, range 0 to 38%) compared to those with normal LVEF (median LGE 11.9%, range 0 to 31%), however, this trend was not statistically significant (p = 0.18). In patients with reduced LVEF (< 60%), there was a subtle trend towards a decreased LVEF with increased LGE extent (r^2 = 0.28, Figure 2).

Conclusion

Myocardial fibrosis is present in most patients with B/D. There is a subtle association between the extent of myocardial fibrosis and impaired LVEF. Myocardial fibrosis in B/D with normal LVEF suggests that fibrosis is either an early disease manifestation or that other factors are required before progression to dilated cardiomyopathy.